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HANSEN'S DISEASE (LEPROSY)

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Leprosy is a chronic, mildly communicable disease of man which primarily affects the skin, mucous membranes, peripheral nerves, eyes, bones and testes due to *Mycobacterium leprae*, an acid fast bacillus related to the agent of tuberculosis.

Hansen's Disease is synonymous to leprosy. Because modern day leprosy is erroneously associated with the biblical leprosy, patients suffering from leprosy are often ostracized and discriminated. A strong movement developed in the Western hemisphere to substitute the name Hansen's disease to leprosy. In many countries the word leprosy is still currently used. However the word leper carries a pejorative connotation and should no longer be used in medical texts.

Epidemiology

Skin to skin transmission has long been suspected to be the main route of transmission. Although bacilli are present in very large number in ulcers, they cannot be found on the unbroken skin. There are a few anecdotal cases of skin transmission: inoculations during surgical procedures and tattooing. Insects have been suspected but careful studies have shown that their role in transmission would only be a minor one (if any).

It seems that the airborne transmission is the more probable route of transmission. Nasal washings from untreated lepromatous cases have from 10,000 to 10,000,000 *M.leprae*. A majority of the lepromatous patients have bacilli in their nasal secretions. The primary infection site may be the respiratory tract or the skin. Aerosols with *M.leprae* have been successful in infecting immunosuppressed mice.

Leprosy is not very communicable. Only 5-10% of the population is susceptible to develop the disease. In the USA the attack rate among close contacts is around 5%. Lepromatous and borderline cases are infectious while indeterminate and tuberculoid are considered as non-infectious. For example, in a study done in 1935 the clinical leprosy attack rates per 1000 person-years were 0.83 for persons with no known contacts with leprosy cases, 1.60 for contacts of non-Lepromatous and 6.23 for contacts of lepromatous cases.

Patients who are adequately treated are no longer infectious a few weeks or months after treatment started. Studies of mothers adequately treated showed that leprosy occurs only among children born prior to the mother's initiation of treatment.

Humans are the only hosts in most countries. In the south central part of the USA (Texas, Louisiana and Mississippi) armadillos are naturally infected by a mycobacterium indistinguishable from *M.leprae*. Prevalence of infection in armadillos range from 4% to 30%. Leprosy has been introduced in Louisiana around the 1850's and the armadillo in 1926. Leprosy is now on the decline in Louisiana. Therefore the "armadillo leprosy" has not played any role in the rise and fall of leprosy in Louisiana. Natural infection were reported from a mangabey monkey (*Cercocebus torquatus atys*) and a chimpanzee (*Pan troglo-*

dytes) from West Africa. The significance of these infections on leprosy transmission is practically negligible.

Age, Sex: Lepromatous leprosy affects males twice as frequently as females but for tuberculoid leprosy the sex ratio is approximately 1:1. In most countries, incidence and prevalence of leprosy are higher in males. The difference is greater in adults than in children. This difference may be due to the higher mobility of males coming more often into contacts with sources of infection, to social taboos and to the usually less thorough examination of the females. Infants rarely have leprosy. The incidence rises in individuals between 10 and 20 and then fall while prevalence rises to a peak between 30 and 50. Children are said to be more susceptible than adults. In families with an index case of leprosy, the incidence of disease is higher among children than among spouses or older siblings.

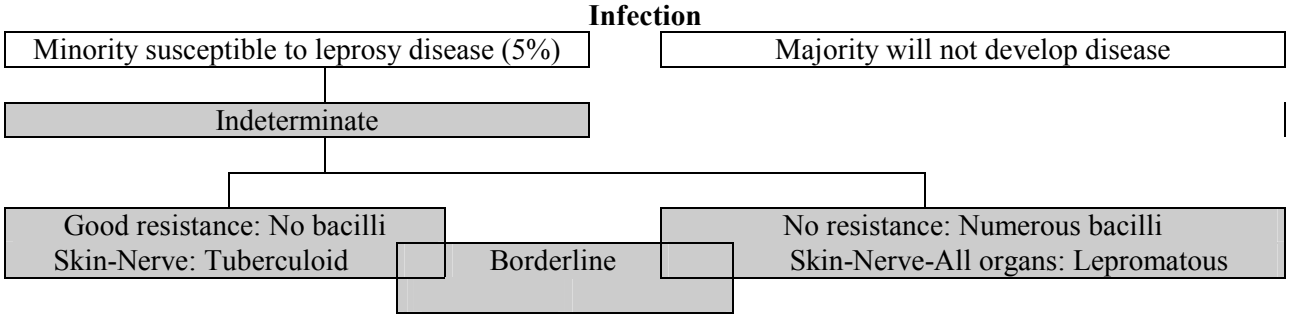
Environmental Factors play a role in transmission. Nowadays leprosy is more prevalent in the warm and humid climates but this has not always been the case. In the last century leprosy was prevalent in countries with cold climate. Crowding, poor sanitation, malnutrition and other environmental conditions seem to favor transmission. M.leprae can remain viable in dried nose secretions for 9 days, in moist soil at room temperature for 46 days. The closer the contacts between susceptible individual and leprosy case, the higher the risk of developing the disease. In a study done in Indonesia, children sharing the same mat as their parents had a much higher incidence of leprosy that children sleeping on a separate mat. Other studies on the effects of crowding have yielded mixed results. It is difficult to differentiate crowding from the other sociologic factors closely associated to it (poor hygiene, poor nutrition).

The disease is not hereditary but a genetic susceptibility may be inherited as shown by the aggregation of cases in some families. Twin data seem to indicate that the concordance of the types of leprosy is higher among pairs of identical twins than among pairs of fraternal twins. HLA linked genes seem to control the type of disease that develops.

Incubation period : The bacillus reproduces at a very slow rate and therefore the incubation period is an average of 3 to 5 years. It is difficult to find out precisely the incubation period because exposure time and degree of exposure are impossible to determine. Few cases were diagnosed in infants less than 1 years, for other cases the incubation may have lasted 20 years or more.

Clinical Description

Infection with M.leprae is detected by lymphocyte transformation test, immunogel diffusion, radioimmunoassay and fluorescent antibody absorption test (FLA-ABS). An ELISA test using a highly specific phenolic glycolipid (PGL) antigen of M.leprae is more sensitive but slightly less specific than the FLA-ABS. Although the results of these tests are diverse, they show that infection with M.leprae is far more common than it is generally accepted: From 5% to 50% of contacts. A positive ELISA is present in 95% of cases 1 or 2 years before the development of clinical signs. There is good evidence that a high proportion of early clinical leprosy (up to 12%) will resolve spontaneously.



Indeterminate type

The indeterminate type is the earliest state of the disease. It consists of a single hypopigmented or erythematous skin lesion which may or may not be insensitive. A skin smear is usually found to be negative. The indeterminate type may heal spontaneously, remain indefinitely at this stage or progress into one of the other types. To confirm a diagnosis of indeterminate leprosy there must be impairment of sensibility (thermal sensibility gets impaired before tactile sensibility). Biopsies are useful to support th clinical diagnosis.

Tuberculoid type

In the tuberculoid type there are few (1 to 3) skin lesions; these are large (3 to 30 cm) macules. The lesions are hypopigmented or erythematous. They have a well defined border and are rough and scaly. The periphery is raised and erythematous or hyperpigmented; the center is flat and hypopigmented.

Anesthesia or hyperesthesia is a major characteristic of these lesions. The sensation of temperature, light touch and pain are impaired in that order. Deep sensation is intact and deep tendon reflexes are normal. Loss of the ability to perspire is an important sign. Peripheral nerve involvement is common. The main nerves involved are the ones in the periphery of the lesions, the ulnar above the elbow, the median at the wrist, the common peroneal at the knee, the posterior tibial at the ankle, the radial at the elbow, the greater auricular in the neck, the sural at the lateral aspect of the ankle, and the superficial branches of the V and VII cranial nerves.

Although tuberculoid Leprosy is a relatively benign disease, severe disabilities may develop.

Lepromatous type

The skin lesions are very numerous small macules. They appear copper colored or erythematous. Their margin is vague and they tend to coalesce. They are symmetrically distributed over the body. They are not necessarily anesthetic. There is a generalized infiltration of the skin, often noticed by palpation rather than by visual inspection. Nodules and papules which are characteristic of lepromatous Leprosy are usually numerous. A common site for the development of nodules are the earlobes. Loss of eyebrows (starting with lateral brows) and loss of eye lashes (madarosis) are a common late signs. Alopecia of the scalp is rare.

As the disease progresses, most organs may become involved: Nasal congestion, epistaxis, laryngeal inflammation, eye lesions, renal involvement, testicular atrophy and secondary gynecomastia. Nerve involvement has a slow onset and becomes more generalized to eventually affect all nerves.

Borderline Type Or Dimorphous Type

Clinically and histologically borderline Leprosy has features of both the lepromatous and tuberculoid types. The majority of leprosy cases are borderline, but for particular purposes, they are often classified as Tuberculoid or Lepromatous (T or L). The word dimorphous is sometimes used instead of borderline.

Skin lesions are a mixture of T or L lesions, with a tendency to be more erythematous. The disease is unstable and may progress towards a more polar form (T or L).

Eye involvement:

The hypoesthesia of the cornea, paralysis of the eyelid muscle, inflammation of the tear duct and lacrosse gland will contribute to the development of lesions due to exposure of the eye. Lagophthalmos is the incomplete closure of palpebral tissue when lids are shut. In lepromatous leprosy, beading of corneal

nerves may be a very early sign. The ciliary body, iris, and cornea are directly involved. As the disease progresses, it will lead to loss of visual acuity, cataract development, glaucoma and eventually blindness.

Nose involvement:

Rhinitis and nose bleeds are common in early lepromatous leprosy. The thick mucous fluid contains large numbers of bacilli and is probably a major source of infection.

Peripheral nerve involvement:

Sensory and motor impairment of the peripheral nerve will lead to the following deformities:

Ulnar nerve	- Clawing of fingers IV and V - Adductor weakness of thumb - Pinch impairment
Median nerve	- Clawing of fingers I and II always associated - Opposition and adduction of /to ulnar) thumb impairment - Abnormal grasp mechanism
Lateral Popliteal	- Loss of dorsiflexion -Loss of eversion
Posterior Tibial	- Clawing of toes

Ulceration;

Ulcers are the result of repeated trauma on the anesthetic areas on the skin. The feet may be injured by poorly fitting shoes. The patient walking barefooted will often injure his feet and be unaware of the injury. The anesthetic hands are also exposed to injury such as burns while smoking or cooking, and traumas while working will ill fitted tools. Secondary infection of the ulcers is common.

Bone involvement:

Secondary infection leads to osteomyelitis and ultimately to bone absorption. Because fingers and toes are the areas most affected, there is partial loss of phalanges occasionally leading to total loss of digits. Direct invasion of bones by *M.leprae* may provoke bone destruction. The nasal bones and hard palate are often involved. Concomitant mucous membrane involvement may result in the perforation of the nasal septum.

Diagnosis

A complete history, physical examination and laboratory tests are the keys to the diagnosis of leprosy. Most leprosy cases either have lived in an endemic area or have been in close contact with a known case of leprosy.

Sensory Changes, chiefly anesthesia, are the most common symptoms reported by the patients.

The key to the diagnosis is a thorough clinical examination**.**
-The total skin area should be examined carefully. Use the brightest available natural light, side lighting may be useful. Examine from close and far distance.
-Light touch can be tested by touching the skin with a wisp of cotton, while the patient's eyes are closed. Some surface areas with thick callous skin are normally less sensitive: elbows, knees, soles, fin-

gers of manual workers. Stroking a hair may cause a sensation. Test tubes with hot and cold water may be used to test temperature perception.
- Palpate for enlarged nerves especially the areas where a superficial nerve may be involved. Watch for wincing during nerve palpation, indicating pain.
- Examine the extremities for contracture and trophic changes.
- Look for a Motor deficit:
 --Facial Nerve - Blink reflex is normally noted when the examiner waves his hand before the eyes.
 --Ulnar, Median, and Radial Nerves - The patient is asked to approximate the tip of the thumb to the tip of the straight little finger with the hand outstretched in the pronated position.
 --Lateral Popliteal Nerve - Patient is asked to dorsiflex the great toe against resistance.
 --Posterior Tibial Nerve- The patient is asked to spread the toes.
-A detailed examination of the eye is necessary. Check the eyebrows, eyelashes, motricity of the eyelids, the cornea, conjunctiva and iris.
-Inspect the mouth and throat.
- Examine the earlobes.

Skin Smear

A skin smear may demonstrate the presence of acid fast bacilli in the skin. If bacilli are located in the deep layers of the skin or in the nerves they will not be found on the smear.

The bacilli in the skin are quantified with the Bacteriological Index (BI). It ranges from 0 to +6 according to the number of bacilli per oil immersion field at microscopic examination. The rate of clearance under treatment is approximately 1 per year. It is a late marker of the antibacterial action of chemotherapy although it is of prime importance in the diagnosis of new cases or relapses.

0	No bacilli	
1+	1 to 10	bacilli per 100 OIF
2+	1 to 10	bacilli per 10 OIF
3+	1 to 10	bacilli per 1 OIF
4+	10 to 100	bacilli per 1 OIF
5+	100 to 1000	bacilli per 1 OIF
6+	Over 1000	bacilli per 1 OIF

The Morphological Index (MI) is the percentage of solidly stained bacilli of normal size and shape. These bacilli are thought to be the viable ones, that is the ones that are alive and may infect someone. There are problems in standardization and reproducibility that cause the index not to be routinely used in practice.

Interpretation of BI and MI: Indeterminate and Tuberculoid leprosy may have a negative or 1+ BI. A nontreated advanced lepromatous may have a 4 to 6+BI with MI from 10 to 50 %. A Borderline may show lower BI and MI. With an efficient treatment the MI will go to 0% rapidly and the BI will generally fall by 1+ every 1 or 2 years. Subjective interpretation by individual technicians vary considerably. Therefore, it is important that the same person perform the microscopic examination to insure consistency in reporting.

A skin smear may be useful for diagnosis: when positive it indicates leprosy but skin smears are used primarily for the surveillance of the patient's bacteriological status.

Technique of the skin smear:
A skin smear may be taken on any suspect lesion, or on the earlobe or eyebrow, the elbow or the knee. This procedure is easy and totally harmless. It can easily be done by physicians, nurses and lab technicians.

-Pinch the skin to reduce blood flow.
-Wipe the area with an alcohol sponge.
-Make a small slit with a sterile scalpel blade or razor blade approximately 5mm long and 2mm deep.
-Wipe away any blood which exudes.
- Scrape the edges of the wound with the blade .
-Spread the materials obtained on a microscopic slide. Obtain as little blood as possible.

Paucibacillary-Multibacillary Cases

Paucibacillary cases are the smear negative I and T cases of Madrid classification (I, TT and BT of Ridley Jopling classification) with a BI=0 at all sites.

Multibacillary cases are L and B cases of Madrid classification (LL, BL, BB and BT of Ridley Jopling classification) with a BI>0. In previous definition, paucibacillary had a BI of 0 or 1 but this was reduced in the latest definitions because of the poor results obtained by therapy in the BT group who had BI of 1.

Skin Biopsy

Examination of a skin biopsy gives the definitive diagnosis and classification of the type of leprosy. The edge of skin lesions or the nodules are considered to be the best sites for obtaining a biopsy. The biopsy is made with a punch or by surgical incision. It should be deep enough to include subcutaneous fat. The specimens are best preserved in neutral buffered formaldehyde solution (a solution containing phosphate buffer and 10 % of formaldehyde 37-40 %).

Inactive Leprosy

The diagnosis of inactive leprosy is important. It means that the disease process has been stopped. The purpose of continued treatment is to consolidate this stage and to prevent later relapses. The patient is no longer infectious. Although physical examination, skin smear and biopsy are all used for the diagnostic, subjective diagnoses are sometimes made depending on clinical and histological impressions. The following criteria are required:
1. Negative skin smears for one year
2. Absence of clinical activity and/or reactions
3. Negative skin biopsy after one year.

Note: Inactivity does NOT mean that all *M. leprae* have been killed in the body. It means that they cannot be found by the usual methods; however, special studies may show the existence of some viable bacilli.

Cultivation & Animal Models

It has not yet been possible to culture the *M.leprae* in vitro. It only multiplies and produces disease in a very limited number of animal species. The nine banded armadillo is the only source of large amounts of *M.leprae* for research and production of vaccine. Inoculation of the normal mouse foot pad is the basic tool used to study *M.leprae* drug resistance and the protective effect of candidate vaccines. Immunodeficient mice (thymectomized, irradiated, bone marrow reconstituted, nude) or neonatally thymectomized rats are also used. Several primates can also be experimentally infected.

Surveillance

Hansen's disease is a condition reportable within 5 working days of diagnosis.

Case Definition

Clinical description

A chronic bacterial disease characterized by the involvement primarily of skin as well as peripheral nerves and the mucosa of the upper airway. Clinical forms of Hansen's disease represent a spectrum reflecting the cellular immune response to *M. leprae*. The following characteristics are typical of the major forms of the disease:

Tuberculoid: one or a few well-demarcated, hypopigmented, and anesthetic skin lesions, frequently with active, spreading edges and a clearing center; peripheral nerve swelling or thickening also may occur
Lepromatous: a number of erythematous papules and nodules or an infiltration of the face, hands, and feet with lesions in a bilateral and symmetrical distribution that progress to thickening of the skin
Borderline (dimorphous): skin lesions characteristic of both the tuberculoid and lepromatous forms
Indeterminate: early lesions, usually hypopigmented macules, without developed tuberculoid or lepromatous features

Laboratory criteria for diagnosis

Demonstration of acid-fast bacilli in skin or dermal nerve, obtained from the full-thickness skin biopsy of a lepromatous lesion

Case classification
Confirmed: a clinically compatible case that is laboratory confirmed

Treatment

Multidrug therapy:

All multibacillary cases (borderline and lepromatous) should be on a multidrug regimen of

- dapsone 100 mg daily (self administered),
- rifampin 600mg (450mg for patients< 35kg) once monthly (under supervision)
- clofazimine 50mg daily (self administered)
- clofazimine 300mg monthly (supervised).

Children 10-14 get dapsone 50mg, rifampin 450mg, clofazimine 50mg. Doses need to be adjusted for children under age of 10

The recommended length of treatment is 2 years or until they become smear negative**.** Then they should be examined at least once a year (clinical examination, skin smear) for 5 years. Never use rifampin alone or rifampin+dapsone without a third bactericidal drug to avoid rifampin resistance.

Monthly supervised ethionamide/prothionamide are not usually recommended and should not be substituted for clofazimine since clofazimine has the added benefit of preventing some reactions.

Paucibacillary cases (indeterminate and tuberculoid) should receive
- dapsone 100mg daily (self administered)

- rifampin 600mg once monthly (supervised) for 6 months.

They should be examined at least once a year for a period of 2 years after completion of treatment.

Reg	Frequency	Drug	Mg	
MB	Daily	DDS	100	⇨ 2 years or longer till smear negative
		CLO	50	
	Monthly	RIF	600	
		CLO	300	
PB	Daily	DDS	100	⇨ 6 months
	Monthly	RIF	600	

-Dapsone Monotherapy is no longer recommended, however some countries still continue using it.

Adult indeterminate and tuberculoid cases were started and maintained on 50 mg of dapsone (DDS) daily, and could be expected to become inactive after 2 years of treatment. Borderline and lepromatous cases were treated with 100mg dapsone daily. Additional treatment lasting 3 years was recommended for I and T, 10 years for B and life for L.

Surveillance of cases is necessary after completion of treatment for 1-detection of relapses and 2-detection of reactions. Patients need to be aware of these two possibilities so that they will voluntarily seek help when either occurs. They must be able to recognize their early sign. PB cases should be examined once a year for 2 years and MB cases for a minimum of 5 years.

Disappearance Of *M.Leprae*:

The lepromatous patient placed under an effective treatment will become non-infectious in a few weeks. However, it usually takes 3 to 10 years before *M. leprae* will completely disappear from a skin smear. Even though the skin shows no evidence of the presence of bacilli, some may persist in the peripheral nerves and internal organs. Premature discontinuation of treatment may allow multiplication of bacilli leading to a relapse.

Single lesion therapy:

A single dose of combination therapy has been used to cure single lesion paucibacillary leprosy
- Rifampin 600 mg
- Ofloxacin 400 mg
- Minocycline 100 mg

A child would take half the adult doses. Blister pack with the drugs are available.

Treatment of leprosy during pregnancy and lactation

Leprosy is exacerbated during pregnancy, so is important that the standard multidrug therapy be continued during pregnancy. The Action Program for Elimination of Leprosy, WHO, Geneva has stated the standard MDT regimens are considered safe, both for the mother and the child, and therefore, should be continued unchanged during pregnancy. A small quantity of antileprosy drugs is excreted through breast milk but the is no report of adverse effects as a result of this except for mild skin discoloration of the infant due to clofazimine. The single dose treatment for patients with single lesion paucibacillary leprosy should be deferred until after delivery.

Treatment of palient with concomitant active tuberculosis.

If the patient has both leprosy and active tuberculosis, it is necessary to treat both infections at the same time. Give the appropriate antituberculosis therapy, addition to the antileprosy multidrug therapy for the type of leprosy in the patient. Rifampicin is common to both regimens and it must be given in the doses required for tuberculosis.

Treatment of patients with concomitant HIV infection

The management of a leprosy patient infected with HIV is the same as that of any other patient. The information available so far indicates that the response of such a patient to MDT is similar to that of any other leprosy patient and management, including treatment reactions, does not require any modifications.

Patient who cannot take rifampicin

Special treatment regimens are required for individual patients, who cannot take rifampicin because allergy or intercurrent diseases, such as chronic hepatitis, or who have been infected with rifampicin-resista leprosy.

In 1997, the WHO Expert Committee on Leprosy recommended the following 24 month regimen for adult patients with multibacillaly leprosy, who cannot take rifampicin:

Length of Treatment 6 months
- Clofazimine 50mg daily
- Ofloxacin 400mg daily
- Minocycline 100mg daily

Followed by an additional 18 months
- clofazimine 50 mg daily
- + ofloxacin 400 mg daily
- or minocycline 100 mg daily

In 1994, the WHO Study Group on Chemotherapy of Leprosy stated that daily administration of 500 mg of clarithromycin can be substituted in the above regimen for either ofloxacin or minocycline during the first six months of treatment of multibacillary patients, who cannot take rifampicin.

Patient who refuses to take clofazimine

Patients with multibacillary leprosy, who refuse to take clofazimine because of skin discoloration, also need a safe and effective alternative treatment. In such patients, clofazimine in the normal 12 month multidrug therapy may be replaced by:

- Ofloxacin, 400 mg daily for 12 or 24 months

Or
- Minocycline, 100 mg daily for 12 or 24 months

In 1997, the WHO Expert Committee on Leprosy also recommended the following alternative 24 month multidrug therapy regimen (3 drugs) for adult patients with multibacillary leprosy, who refuse to take clofazimine:

Rifampicin, 600 mg once a month for 24 months
Ofloxacin, 400 mg once a month for 24 months
AND minocycline, 100 mg once a month for 24 months

Patient who cannot take dapsone

If dapsone produces severe toxic effects in any leprosy patient, either with paucibacillary or multibacillary leprosy, dapsone must be immediately stopped. No further modification of the regimen is required for patients with multibacillary leprosy. However, clofazimine in the dosage employed in the standard multidrug therapy for multibacillary leprosy should be substituted for dapsone in the regimen for paucibacillary leprosy for a period of 6 months.

Reactions

Reactions are the chief complications occurring during treatment of the disease, they are a major source of morbidity. They are 2 types of reactions:

1-Reversal Reactions or upgrade or Type I reactions. These occur in a wide variety of patients, from BT to subpolar L, they are more common among BL patients. They are due to a delayed hypersensitivity reaction. They may occur before initiation of therapy or during the first 2 years of treatment. This type of reaction has become more common with multidrug therapy. The diagnosis is primarily clinical. The major signs are:
-Fever and constitutional symptoms may occur but are not prominent.
-Inflammation of skin lesions: they take a dark red or purple hue, they become rougher, more inflamed and are edematous, but they are not tender. Spontaneous ulceration is rare.
-Neuritis: inflammation of the nerves which may give rise to a sensory or motor deficit. It is of utmost importance that cases of neuritis be treated at a very early stage. Check for motor deficit, sensory deficit, pain and tenderness. Early diagnosis of neuritis and of quiet nerve paralysis is made much easier if careful records of cutaneous sensibility (sensory maps) and records of muscular strength of hand and foot muscle are kept regularly.
-Quiet nerve paralysis may occur without overt neuritis. This condition may easily be missed in the early stages when recovery is possible and is recognized only after serious and irreversible damage is done. The onset of nerve damage is insidious. The patient may have some difficulties describing the subjective sensory disturbances and the motor weakness. The administration of steroids at much lower doses than for the treatment of severe neuritis is indicated to prevent the establishment of nerve trunk paralysis. In patients with enlarged nerve trunks, get a good history of paresthesias, make a good sensory map and carefully test for muscular strength.
-Edema
-Scarring and permanent paralysis may result from untreated reactions.

2-Erythema Nodosum Leprosum or Type II reaction. This reaction occurs in L or BL patients. It has its onset within 2 years after initiation of therapy but may also start prior to treatment. It is due to an Arthus phenomenon. It was quite common with dapsone monotherapy but is less frequent with multidrug therapy. The major signs are:
-Painful, tender erythematous dermal and subcutaneous nodules occur on the face, and occasionally all over the body. They are bright pink and evolve to a deep red. In the Bantus the subcutaneous nodules predominate. Vesicules, pustules and bullae are less common. There is a brawny induration of the subcutaneous tissues, particularly in the postero-lateral areas of the arms and thighs.
-The red leg phenomenon is an erythema, edema and tenderness simulating cellulitis occurring on the leg (or rarely forearm).
-The eye may be inflamed and painful. Several inner organs may be inflamed: hepatomegaly, orchitis.
-Peripheral nerve involvement result in neuritis and paralysis.
-There may be swelling of the joints, peripheral edema and lymphadenitis.
-Severe constitutional symptoms as fever, chills, malaise, anorexia and weight loss.
-Leukocytosis around 15,000/mm^3 and neutrophilia are present in 80% of ENL. A normocytic, normochromic anemia is common in L cases and in ENL cases as well. During the ENL some patients experience an abrupt fall in hematocrit.

3-The Lucio Reaction is restricted to patients from Mexico, Central America and Cuba. It occurs only in patients with diffuse non-nodular LL form (Latapi's lepromatosis). It is characterized by stellate hemorrhagic infarcts which heal with crusting or ulceration and scar formation.

Treatment of reaction:

Mild reactions require little or no treatment. Aspirin may be helpful. Moderate to severe reactions should be hospitalized. Thalidomide , corticosteroids and clofazimine are used to treat these more severe reactions.

Treatment of reversal reactions:

-Steroids are specially indicated in case of acute neuritis. The usual dose is a single 30-60mg am for one week, if there is no improvement the dose should be increased up to the level where the symptoms are alleviated. When a good response has been maintained for 3 weeks, the dose may be tapered in 5mg increments weekly until 20mg daily is reached before switching to alternate days.
-Clofazimine is too slow to act as an effective anti-inflammatory agent in the beginning of treatment of the reversal reaction. It is introduced later as a steroid sparing drug at doses of 200mg daily. If a nerve is involved in a fibrous canal and response to medical management is not prompt, neurolysis should be considered.

Treatment of ENL:

-Thalidomide is an excellent drug used to treat severe ENL reactions. Its use is severely controlled because of its teratogenic effects. Other side effects are minor: sleepiness, visual disturbances and malaise. Most patients respond well to 100mg at bedtime but some require more. Patients not responding to 200mg require steroids or clofazimine. If thalidomide is contra-indicated, steroids should be used.
-Steroids are used to control the acute phase of the reaction.
-Clofazimine is used for long-term control of reactions and steroid sparing.

Drug resistance:

M. leprae may develop resistance to the drugs. The use of Dapsone began around 1948, the first cases of resistance were suspected in the early 60's and by 1975 it became obvious that dapsone resistance was a major problem. Sulfone resistance develops in perhaps 10 % of lepromatous leprosy. It takes an average of 10 to 15 years to develop. It is more likely that patients taking low doses or frequently interrupting the treatment will develop drug resistance. When rifampin is used alone, resistance may develop within a few years. To date, there is no evidence of developing resistance to clofazimine.

Suspect resistance or non-compliance when:
-The patient's condition does not improve over a period of l year, assuming other complications do not develop which could confuse the picture.
-There is no reduction in the patient's Morphological Index (MI) and Bacteriological Index (BI) over 1 or 2 years.

The mouse foot pad studies for the drug resistance allow confirmation of drug resistance. These are performed only in very specialized centers. Send a sterile , dry, 4 to 5 mm punch biopsy of a skin area with a BI of 4+ or more. Do not put any preservative in the sterile tube where the biopsy is. Pack in wet ice if delays in transportation are not too long.

Management of resistant leprosy:

Rifampin, clofazimine, ethiomanide are often used in combined therapy for treatment of DDS resistant cases. Such cases are difficult to treat and supervise.

Relapses

Ideally relapse rates should be computed by using the number of person-years of actual observation as a denominator and not the number of person who were negative at the beginning of the study. Relapse rates depend on the regularity of treatment, chiefly during the smear positive period.

For MB leprosy treated with dapsone monotherapy, they range from 0.5% to 2.5% person-years according to the regularity of treatment. The cumulative probability show a sharp increase during the first 5 years to reach a plateau by 10 years, by then 10 to 25% of patients would have relapsed.

For MB leprosy treated with multidrug therapy 60-70% of patients are negative within 6 months of treatment and 95% by 1 year. Relapse rates are in the order of 1-3% person-years of follow up.

Leprosy Control

The main objectives of a leprosy control program are:
1-To interrupt transmission of the infection, reducing the incidence of the disease so that it no longer constitute a public health problem.
2-To find new cases that are symptomatic or before symptoms develop.
3-To follow up all known cases and ascertain that they receive proper medical care and take regular treatment.
4-To prevent the development of associated deformities
5-To educate the patient, his family, the health professional, and the public.

In recent years, leprosy control programs have had to deal with increasing secondary and primary resistance of *M.leprae* to dapsone. The main strategy for control remains early detection of cases and chemotherapy.

Identify Suspects is the primary goal of case finding. The health practitioner comes in contact with many persons during the workday. They should be aware of leprosy and always suspect it if working in an endemic area. Suspects should be examined promptly by a person (physician, nurse) experienced in the diagnosis of leprosy.

ALL new cases should be treated, even paucibacillary cases. Although a proportion of these cases can heal without therapy, there is no way to tell which one will heal and which one will get worse.

Following the diagnosis of a new case, it is important to start a Case Investigation. The purpose of this investigation is to provide epidemiological information, baseline clinical information on the patient, and contact information for follow up. Collect information on the patient's identity, address, ethnic group, birth, education and employment. The places of residence should be investigated with great care. The information provided is essential to determine where the patient was contaminated, clinical information at the time of diagnosis. It is important to note the clinical type of leprosy since it will determine the length of treatment and the length of follow up of contacts, names of contacts and possible source of infection. The source of infection is considered as anyone having leprosy that came into contact with the patient. It is impossible to ascertain whether this person was actually the source of infection. The most important contacts are first the bedroom contacts, then the household contacts, then relatives, friends, co-workers not living in the household, but only if there is frequent contact. Judgment is necessary in determining who should be listed as a contact.

Programs should promote Self Detection through health education, as self reported patients generally are better compliers. Where multidrug therapy has been successfully implemented, self reporting has been improved.

Bacteriological Examination is highly relevant to leprosy control programs. The quality of smears and of microscopy is often the weakest link in most leprosy control programs. It is essential to train personnel in proper collection procedures for smears and to organize an efficient system for processing the smears.

Case Notification

New cases are reportable.

Containment

Isolation

Isolation was used for many centuries as the only way to prevent spread of the disease. It has not proven very efficient. Patients knowing they would be isolated for a long period of time avoided medical care and attempted to hide their disease. Although they were isolated when the diagnosis became obvious, it was usually very late. Most contacts were already contaminated. Infectious cases become noninfectious within a few weeks or months following the initiation of treatment. Isolation of the patient is no longer necessary. Common sense and hygienic precautions with regards to an infectious case are sufficient.

Hospitalization

Following the diagnosis of a new case, hospitalization is not systematically recommended before starting treatment. The major medical indication for hospitalization is in the event of a REACTION to drug therapy. Severe reactions may lead to severe disabilities and death. It is to the patient's benefit to be hospitalized when a reaction occurs. Cases that are diagnosed early with minimum signs of leprosy do not need to be hospitalized and may begin treatment as outpatients.

Follow Up Of Known Cases

One must understand the public's prejudices toward leprosy. Persons with leprosy have been unfairly and irrationally ostracized and sequestrated for many years. An understandable and common reaction from patients is to refuse any control from anyone. A great deal of tact is necessary in dealing with these cases.

The health practitioner should record the successive addresses of the case; due to the usual length of treatment and need for follow-up. The patients should be referred when they move out of the jurisdiction, to the appropriate health center serving the patient's new location.

New leprosy cases need to be seen at least every month for the time of drug administration. The purpose of the follow up visits are:
1-to assess the patient's condition and his needs for specialized care
2-to check on the regularity of treatment and for the presence of any signs of reaction
3-To administer directly supervised drugs and renew other drugs

Compliance is a major problem in leprosy control. The old regimen of monotherapy with dapsone was partly unacceptable because of the requirements for extremely long treatment: 3, 5 years or lifetime treatment. With multiple drug therapy the length of treatment have been considerably shortened but are still very long in comparison with treatment for other diseases: 6 months, 2 years or longer. The magnitude of the non-compliance is seldom assessed. Rates of defaulters vary according to studies and defini-

tions used: around 10% per year, at the end of 5 years of unsupervised monotherapy it was not unusual to have only 25 to 50% of patients still attending clinics. Mere attendance at the clinic does not mean that the patient takes faithfully his drugs. Urine controls of drug intake showed that 50 to 70% only of patients attending clinics do take their drugs regularly. With multiple drug therapy and monthly supervised drug administration, compliance improved slightly. Patients seem to prefer the new regimen and make an effort to be more regular. Patients can see faster results and are told that they will be released from treatment within near future, therefore their motivation is higher. Monitoring patient compliance is essential. Attendance at clinic appointments is essential however it is not sufficient. To monitor the compliance at home, patient interview are inadequate. Even pill counts may overestimate compliance. A urine test is probably the best option for monitoring drug intake.

Contact Examination

The health practitioner should carry out the identification and examination of close contacts. Household contacts are considered to be at highest risk. They are all persons who have lived in a living unit with a patient for at least one month between the onset of symptoms and the time the patient is considered non-contagious. A health practitioner or a nurse trained for this procedure may perform the screening examination and arrange for this patient to be examined by a physician or a more experienced person.

Guideline For Screening Contacts: select a suitable area where the patient will not be embarrassed by onlookers for performing the examination.
1-Skin Examination: Begin the examination at the head and proceed down to the extremities.
-Examine the entire skin surface for any lesions (patches, nodules, ulcers...)
-Check for thinning or absence of eyebrows.
-Examine earlobes for nodules and enlargement.
2-Neurological Examination:
-Examine the skin lesions, the medial parts of the hands and the feet for decreased sensation. Use a wisp of cotton and have the patient point to the area that was touched.
-Check the ulnar groove for enlarged ulnar nerve
-Check the popliteal area for enlarged peroneal nerve
-Check for decreased motor function ability in the hand: ability to oppose thumb to each finger (medial nerve), ability to spread fingers apart and resist any opposition.

If no lesions resembling leprosy are found, consider the patient Non-Suspect and advise him on leprosy signs and the necessity for regular follow up examinations. If any suspect lesion was found:
-Obtain a skin smear on all suspect skin lesions and on both earlobes.
-Arrange for the patient to be seen by a physician or nurse with experience in leprosy.

Frequency Of Contact Examination:The frequency of contact examination is based on the infectiousness of the index case.
-Lepromatous & borderline case: Screen contacts once a year for 10 years after diagnosis of case.
-Tuberculoid & indeterminate case: Screen contacts once a year for 5 years after diagnosis of case.
The following are factors to be taken into consideration: 1-Degree of closeness of contact: A close bedroom contact needs to be followed longer than an occasional visitor; 2-Infectiousness of the case: Contacts of patients with high bacteriological and morphological index need to be followed for a longer time; 3-Age of contacts: Young children are thought to be more susceptible to leprosy and therefore should be followed for a longer time; 4-Regularity of treatment of index case: An index case that does not take medication regularly will continuously expose his contacts; 5-Development of resistance : Contacts of drug resistant cases should be followed for 10 years following the initiation of an effective treatment.

Chemoprophylaxis

Dapsone and Acedapsone were effective in the chemoprophylaxis against leprosy in studies carried out in Micronesia, China and India. Acedapsone (150-225mg /10weeks) was 55% effective in India, 95% in Micronesia, Dapsone (1-4mg/kg per week) 35-75%. Prophylactic treatment had to be given at least 3 years or until the index case became negative. The maximum benefits was in children exposed to multibacillary cases. The project in Micronesia failed because of inadequate control of lepromatous cases who relapsed and infected children after the cessation of trial.

Education

The success of an education program depend on the quality of the communication between the practitioner and the patient. The patient population reflects the cultural diversity of the country. Patients and practitioner employ different cognitive systems for understanding sickness resulting from differences in cultural and educational backgrounds.

Practitioners are well aware that leprosy has strong social implications. In many cultures leprosy cases are stigmatized, but this is not the case in all cultures.

It is useful to elicit from patients their explanatory model for the sickness. This would give valuable information about the cultural context of experiencing sickness and shed some light about the potential conflicts between patient and practitioner's perspectives. Areas to be explored are: 1-etiology, 2-time and mode of onset of symptoms, 3-pathophysiology, 4-course of sickness, 5-treatment.

Many patients embrace differing explanatory concepts using using biomedical and traditional concepts as well. The germ theory of disease and the natural consequence of killing the organisms as a mean to obtain cure originated in the western culture and is not universally used. It does not provide a satisfactory answer to many questions. Why does this germ affect only some people and not the others. Why me ? Bad luck, demerit or some circumstantial event are often cited as the cause of disease. A survey on the knowledge about the evolution of the disease done in the USA showed that very little factual information was known about leprosy. The popular view is one of a fantasy of extremely severe and debilitating illness. Treatments are often started and discontinued according to the perception of benefits and losses. Patients frequently change practitioners and treatment throughout the course of the illness. Different events may not be seen by the patient as the many possible outcomes of a single disease. Ulcers, macules, loss of sensation, claw hand, shortening of the hands are seen as different sicknesses. Patient education should stress the relationship between symptoms and repeatedly emphasize the need for prolonged therapy. A good knowledge of the local traditional illness beliefs may provide some culturally meaningful explanations that can be used. Modern chemotherapy seldom wins over patients by its own merits.

Educate The Patient: The health practitioner assumes responsibility for the initial counseling and education of the patient and his family. The health practitioner should ascertain the patient's level of knowledge regarding leprosy in addition to thoughts and feelings about having the disease. Misconceptions should be corrected. The anxiety level of the patient may limit the patient's ability to engage in effective teaching during the first interview. The basic facts, however, should be presented. It is recommended that several interviews be conducted in order to repeat and reinforce the teaching. False reassurance contributes to the development of a non-therapeutic relationship and is unfair to the patient. It takes time for a practitioner-patient relationship to develop. Whenever possible, the same nurse should provide care over a period of time. Common to patients with leprosy is the fear that the diagnosis will become known by others. The practitioner must respect this desire for confidentiality.

Points to emphasize to the patients are:
-the necessity for regular treatment and surveillance
-the good prognosis of Leprosy treated regularly
-the possibility of reactions, their early signs and the necessity to seek medical care for reactions.

-In the case of existing disabilities, the patient should be taught to cope with them. Special information to cover are: 1-Care of insensitive hands, 2-care of insensitive feet, 3-care of the eyes.

Educate The Family: The family and close contacts of the case have been exposed to the leprosy bacilli and should be examined regularly for early signs of leprosy. The nurse should:
-emphasize the purpose of these systematic examinations, explain their frequency;
-teach the contacts how to check for early signs of leprosy; change in skin color, change in skin sensitivity, nodules in any part of the body, change in motor function;
-stress the importance of the contact to have any suspect lesions checked by a health person;
-discuss the transmission of Leprosy, the immunity of the majority of people, the incubation and any other relevant epidemiological information;
-answer any questions the contact may have regarding leprosy.
-assist the contact to adjust and accept the need of regular check ups.

The family plays an important role in the patient's care and progress. They should be taught basic facts in the care and evolution of leprosy.

Educate The Public: The public health practitioner has a prime responsibility in public and individual education. It is desirable that the community have a wholesome attitude toward leprosy as a communicable disease. The attitude of the community toward the patient largely determines the ease or difficulty with which the patient returns to his original environment and job. The goal of the public education should be to remove all erroneous stigma and dishonor from leprosy patients.

Educate Health Professionals: Many health professionals hold similar misconceptions about leprosy. Opportunities should be used to correct these misconceptions and to provide education regarding leprosy.

Social Problems Of Leprosy

The importance of social and cultural factors in leprosy control and the serious implications of these factors on the leprosy patients are now well recognized. The ancient practice of isolation has helped perpetuate the stigma of leprosy in many countries. Since cases are treated at home, it become obvious that customs, culture, social attitudes and restrictive laws of the past have a great impact on leprosy control and on the well being of patients. Therefore it is important that leprosy workers become involved in the efforts to improve the social and economic conditions of patients through health education and other activities.

Hospital precaution and isolation: Standard precautions